Case report

Tetraparesis as clinical correlate of subacute cervical flexion myelopathy

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Context: We report the case of a 20-year-old woman who underwent tracheal resection with postoperative chin-to-chest suture for 10 days, presenting with severe tetraparesis at our institution. Similar cases have been reported previously, however, not yet in the pathophysiological context of chronic cervical flexion myelopathy (Hirayama syndrome).

Findings: Extensive myelopathy at cervical level is the consequence of the fixed cervical spine position due to chin-to-chest suture. Predominantly affected young individuals (age range from 20–25 years) without evidence of degenerated spine disease suggest a similar mechanism as described in Hirayama syndrome—displacement of the dura with consecutive compression of the spinal cord vasculature.

Conclusions: Subacute flexion myelopathy represents a serious complication of operative/postoperative fixed cervical spine positions, warranting particular attention by respective surgeons.

Keywords: Tetraplegia, Chin-to-chest suture, Hirayama, Cervical flexion myelopathy

Introduction

Hirayama syndrome, a slowly progressing juvenile atrophy found predominantly in Asian countries, is thought to be caused by a chronic cervical flexion myelopathy. Here, we report the case of a tetraparesis due to a subacute cervical flexion myelopathy.

Case report

In June 2013, a 20-year-old woman presenting with increasing dyspnea underwent surgical resection of a subglottic tracheal stenosis with consecutive end-to-end anastomosis. Postoperatively, a leakage at the anastomosis required surgical re-anastomosis. In order to reduce tension at the anastomosis, a chin-to-chest suture was performed and the patient was sedated and ventilated for 10 days. After sedation was discontinued, the awake and alert patient presented with a motor complete tetraplegia with deep anal sensation being preserved, reduced sphincter tone and no volitional sphincter contraction (according to the ASIA impairment scale level B; AIS-B) without any voluntary arm or leg movements, and reduced light touch and pinprick sensation beginning at C5 level. Bladder control was absent. MRI of the cervical spine revealed longitudinally extending, dorsally accentuated signal changes from C3 to Th1 with swelling of the spinal cord. No contrast agent uptake within the spinal cord was present. Interestingly, at corresponding levels signal changes were also observed in paraspinal muscles bilaterally with homogeneous contrast enhancement within these muscles (Figs. 1A–D). Imaging findings were interpreted as myelopathy either due to ischemia or venous congestion. The CSF analysis was unremarkable. Neither steroid medication nor surgical interventions were applied. Over the course of several months a comprehensive rehabilitation therapy program allowed the patient to improve sensorimotor function. At discharge the patient regained complete motor function in the upper extremities, and lower extremity motor scores improved to on average 3/5 with persistent hypoesthesia and reduced vibration; however, the patient was unable to stand or walk unsupported. Deep tendon reflexes were brisk with bilateral pyramidal signs and a spastic tone in the lower extremities. Neurogenic bladder dysfunction persisted. Follow-up MRI 14 weeks later showed a reduction of cord swelling with hyperintense signal changes now confined to the

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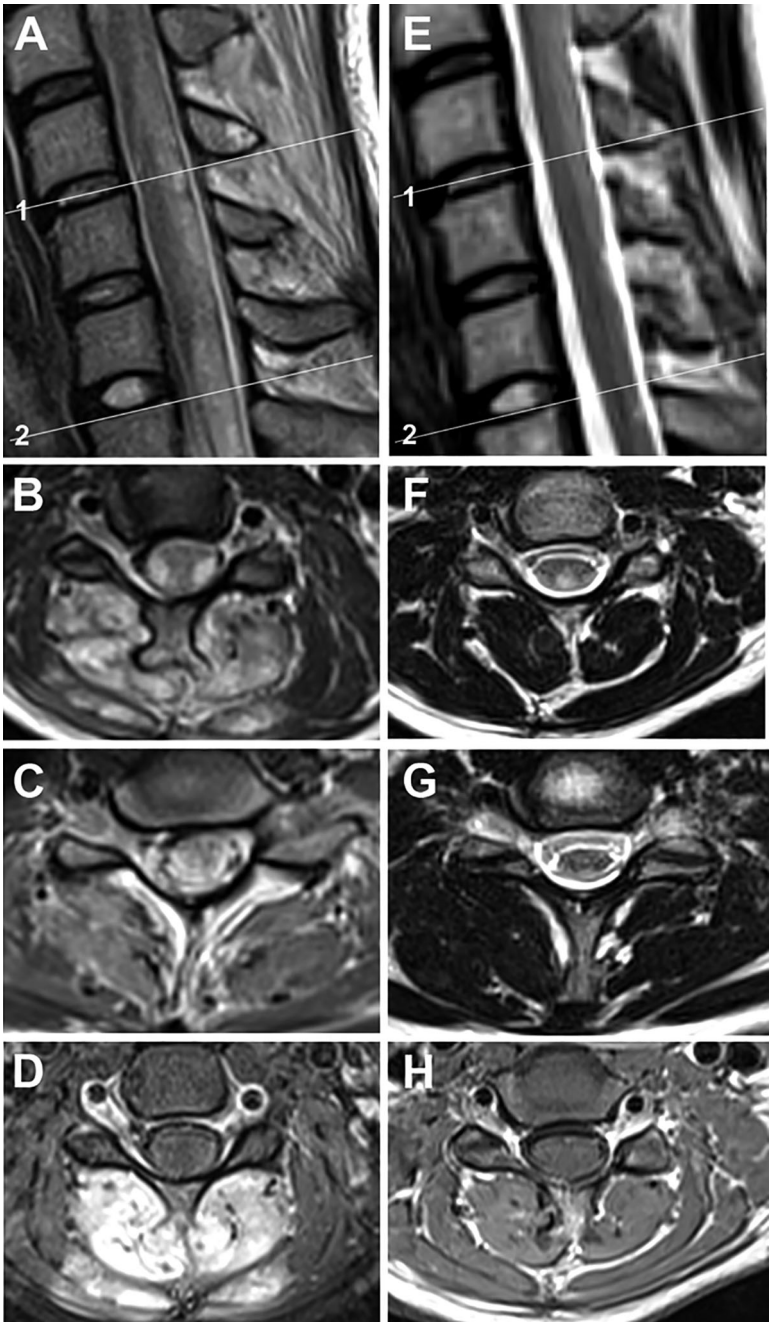

Figure 1 (A) Sagittal T2-weighted 1.5 Tesla MR image demonstrates swelling of the spinal cord and dorsally pronounced hyperintense signal changes extending from C3 to Th1. (B) The axial T2-weighted sequence at C4/5 level (corresponding to line 1 in A) shows the increased signal dorso-laterally, mostly affecting the lateral columns including the corticospinal tract and sparing most of the gray matter and the dorsal columns. Swelling and homogeneous T2-weighted hyperintense signal of the spinalis cervicis, multifidus, semispinalis capitis, and semispinalis cervicis muscles on both sides is depicted. (C) At C6/7 level an axial T2-weighted image (corresponding to line 2 in A) shows a more diffuse pattern of hyperintense signal changes in the spinal cord with less pronounced signal changes in paraspinal muscles compared to B. (D) The axial fat-suppressed contrast-enhanced T1-weighted sequence at C4 level demonstrates the homogeneous contrast enhancement within these muscles. Of note, there is no contrast enhancement in the affected spinal cord. (E) The sagittal T2-weighted 14 weeks later shows reduced swelling of the spinal cord and a decrease in T2-weighted hyperintense changes. (F) Axial T2-weighted image at C4/5 level (corresponding to line 1 in E) shows hyperintense signal changes confined to the dorsal columns. Edema-like signal changes within the paraspinal muscles disappear and respective muscles are moderately atrophied (G) The axial T2-weighted image at C6/7 level (corresponding to line 2 in E) depicts areas of low signal intensity within the central spinal cord indicative of blood degeneration products. (H) In the axial contrast-enhanced T1-weighted sequence at C4 level 14 weeks later contrast enhancement within paraspinal muscles is hardly detectable. The size of affected muscles is reduced.

dorsal columns together with signal reduction in the dorsolateral white matter indicative of a hemorrhagic component. The edema-like signal changes and the contrast-enhancement within the aforementioned paraspinal muscles disappeared, however, the muscles showed moderate atrophy (Figs. 1E–H).

Discussion

We propose to term the observed spinal cord injury as subacute cervical flexion myelopathy. Besides the presented case, a number of other cases of spinal cord injury due to protracted fixed cervical spine positions—predominantly in young individuals (range 18–48 years of age; median 24 years of age)—have been reported in the course of surgeries requiring a flexed cervical spine position, unconsciousness due to medication overdose or after an assault forcing the victim into a flexed cervical spine position for a prolonged period of time.[1–5]

In the literature, the term cervical flexion myelopathy is reserved for a chronic disease condition also known as Hirayama syndrome. Typically, young individuals between 15 and 25 years of age present with a slowly progressing amyotrophy in the upper extremities, which is in most instances self-limiting within a few years. Pathophysiologically, a combined mechanical/ischemic mechanism is discussed. Insufficient growth of the dura relative to the spine during adolescence causes forward displacement of the dura in flexed positions of the cervical spine.[6] The subsequent compression of the spinal cord against the vertebral bodies compromises microcirculation.[7] In the slowly progressing Hirayama syndrome, damage to the cord parenchyma is confined to the anterior horn of the spinal cord gray matter with consecutive flaccid paresis in the upper extremities.[8]

The flexed cervical spine position in the present case with consecutive ventral shift of the dura likely represents the identical trigger as being proposed for Hirayama syndrome. We cannot provide direct evidence such as flexion MRI to support our hypothesis. However, cervical spine flexion does not compromise the osseous diameter of the spinal canal. Therefore, compression of the complete cord by these means is unlikely.[9] Alternatively, neuroforaminal narrowing could compress entering segmental arteries or exiting veins. However, the collateral network of the spinal cord is so extensive that the occlusion of one or two segmental arteries would be highly unlikely to cause a spinovascular problem. Differences in terms of the duration and recurrence of the flexed spine position in our case and Hirayama syndrome can account for differences in terms of neurological presentation and diagnostic findings. In case of Hirayama syndrome the hypermobile dura compresses the cord microcirculation repeatedly for a short duration.[7] As a consequence only highly susceptible neural cells within the spinal cord, namely motoneurons in the ventral horn are affected with isolated lower motoneuron signs and minimal MRI changes. In contrast, in our case or any of the previously reported cases of subacute cervical flexion myelopathy the shift of the dura over longer periods of time (hours to days) may compromise larger caliber blood vessels (posterior spinal artery, epidural veins) causing more extensive circulatory problems affecting the white matter structures predominantly in the dorsal half of the spinal cord. This is paralleled clinically by upper motoneuron signs and prominent MRI findings in respective regions. In case of vascular compression through the hypermobile dura arterial or venous circulation may become affected. In the present case, underlying venous hypertension is supported by axial distribution of signal changes not exactly confined to the supply area of the anterior spinal artery or posterior spinal artery (Fig. 1). Concomitant hemorrhage, which is associated with a less favorable outcome in traumatic spinal cord injury, may have contributed to the poor recovery in the present case. Furthermore, the concept of an underlying venous congestion is supported by the observed changes in paraspinal muscles, which most likely reflect myonecrosis in the context of a flexion induced compartment syndrome. Venous congestion within paraspinal muscles can be induced by compression of the posterior external venous plexus communicating with the internal venous plexus and the veins draining the spinal cord. Such circulatory causes of compartment syndromes have been reported for thigh muscles.[10]

This case should raise the awareness to check the necessity for surgeries requiring similar intra- or postoperative flexed cervical spine positions very carefully, particularly in young individuals, who are predominantly affected by subacute cervical flexion myelopathy.[1–5] In cases, where respective positions cannot be avoided, intra- and/or postoperative neuromonitoring should be considered to detect spinal cord dysfunction before irreversible damage to neural tissue occurs.

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References

1 Kaye KL, Ramsay D, Young GB. Cervical flexion myelopathy after valproic acid overdose. Spine (Phila Pa 1976) 2001;26(19): 459–61.
2 Haisa T, Kondo T. Midcervical flexion myelopathy after posterior fossa surgery in the sitting position: case report. Neurosurgery 1996;38(4):819–21.
3 Levy LM. An unusual case of flexion injury of the cervical spine. Surg Neurol 1982;17(4):255–9.
4 Dominguez J, Rivas JJ, Lobato RD, Diaz V, Larru E. Irreversible tetraplegia after tracheal resection. Ann Thorac Surg. 1996;62(1): 278–80.
5 Windfuhr JP, Dulks A. Spinal cord infarction following cricotracheal resection. Int J Pediatr Otorhinolaryngol 2010;74(9):1085–8.
6 Lehman VT, Luetmer PH, Sorenson EJ, Carter RE, Gupta V, Fletcher GP, *et al.*. Cervical spine MR imaging findings of patients with Hirayama disease in North America: a multisite study. AJNR Am J Neuroradiol 2013;34(2):451–6.
7 Elsheikh B, Kissel JT, Christoforidis G, Wicklund M, Kehagias DT, Chiocca EA, *et al.* Spinal angiography and epidural venography in juvenile muscular atrophy of the distal arm "Hirayama disease". Muscle Nerve 2009;40(2):206–12.
8 Hirayama K, Tokumaru Y. Cervical dural sac and spinal cord in juvenile muscular atrophy of distal upper extremity. Neurology 2000;54(10):1922–6.
9 Muhle C, Wiskirchen J, Weinert D, Falliner A, Wesner F, Brinkmann G, *et al.* Biomechanical aspects of the subarachnoid space and cervical cord in healthy individuals examined with kinematic magnetic resonance imaging. Spine (Phila Pa 1976) 1998; 23(5):556–67.
10 Best IM, Bumpers HL. Thigh compartment syndrome after acute ischemia. Am Surg 2002;68(11):996–8.